Case 1: Radiation induced fibrosarcoma following treatment ofesthesioneuroblastoma

A 59-year-old female presented with frontal headaches, vision changes, proptosis, and fatigue five years following resection and adjuvant chemoradiation of a Kadish stage C-Hyams grade II/IV esthesioneuroblastoma.

**Imaging:**
An MRI was performed that demonstrated an enhancing 3 x 2.2 x 2.8 cm lesion in the right ethmoid cavity extending into the right orbit and along the anterior cranial fossa (Figure 1).

**Treatment:**
The patient underwent a bifrontal craniotomy, right orbital exenteration, craniofacial resection with ethmoidectomy and sphenoidotomy, and radial forearm free flap reconstruction.

**Pathology:**
Pathology revealed a grade 3/3 adult-type fibrosarcoma with invasion into the bone, necrosis, and positive margins. Stains were negative for s100, SMA, desmin, Muc4, and c-myc (Figure 2).

**Outcome:**
Her post-operative course was complicated by areas of wound dehiscence requiring multiple debridements. Six months following surgery, the patient lost vision in her remaining eye. Repeat imaging revealed diffuse leptomeningeal metastasis in the right parietal dura and sella region. Palliative chemotherapy was initiated.

**Discussion**
Radiation induced sarcomas of the skull base are a rare entity that tend to be higher grade and more aggressive than their primary tumor counterparts. The median latency period between radiation and tumor diagnosis is 10-12 years with a steep increase in incidence at 5-13 years. However, in cases where these tumors may occur more than 20 years after radiation, 1,3 as with these two patients, most present with headache or vision change. The most common radiation induced sarcoma types are fibrosarcomas (76%), osteosarcomas (10%), and malignant fibrous histiocytomas (6%). The location of the tumor within the skull base is a particularly challenging problem as complete resection is generally not possible due to involvement of vital structures. As a result, the local recurrence rate is high at approximately 86% and the majority of patients die within the first 6 months after diagnosis.

**Conclusions**
Radiation induced skull base sarcomas pose a significant risk to patients successfully treated with radiation and have an especially poor prognosis. It is important to recognize the risk of this late complication in all patients receiving radiation to the skull base, regardless of primary tumor type. Long-term follow up of post-radiation patients is essential and any symptoms consistent with a radiation –induced malignancy should prompt immediate work up.